

of congenital heart disease (CHD). To assess the impact of FE, we analyzed data from the Metropolitan Atlanta Congenital Defects Program (MACDP), a population-based surveillance system for serious birth defects. Between January 1, 1990 and December 30, 1994, MACDP ascertained 1491 cases of CHD, for a birth prevalence of 8 per 1000 live births. In addition, we ascertained FE-diagnosed congenital heart defects to residents of the same population.

Of the 1491 cases of CHD, we found 70 cases where cardiac defects were diagnosed using FE (5%). FE diagnosis increased from less than 3% in 1990 to almost 10% in 1993. In addition, the types of heart defects diagnosed by FE varied considerably from the general birth population. Diagnoses using FE were made on 23.1% of cases of Hypoplastic Left Heart Syndrome, 15.1% of all Atrio-Ventricular Septal Defects, 13.1% of all cases of Tetralogy of Fallot, 6.5% of cases of Hypoplastic Right Heart Syndrome, and 5.8% of all cases of Transposition of the Great Arteries. The death rate for liveborn FE cases was 37.1% compared to 10.8% of infants diagnosed postnatally. Fetal demise was high among the prenatally diagnosed group with 10.0% being stillborn.

In conclusion, prenatally diagnosed cardiac defects are more severe and have a poorer outcome than defects diagnosed postnatally. FE will continue to have a profound impact on the practice of pediatric cardiology and the ability to conduct epidemiologic surveillance of cardiac defects in the population.

10:45

746-2 Etiology and Outcome of Fetal Myocardial Diseases

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The aim of this study was to evaluate our clinical experience with myocardial disease (MD) diagnosed in utero. Data from 860 patients (pts), 1374 fetal echocardiograms performed in our institution (1983–1995) were reviewed. Thirty one cases (3.6%) had MD.

Echocardiographic data including chamber size, wall thickness, cardiac areas, parameters of systolic and diastolic function, were analyzed and compared to 91 normal fetuses, according to their gestational age (GA). Postnatal echocardiographic data and pathology, when available, was also reviewed.

Results: 1) 24/31 pts (81%) were fetuses of diabetic mothers with hypertrophic cardiomyopathy (HCMO) without outflow obstruction. HCMO was recognized between 31–38 wks GA, mean 34.5 wks. 6/24 pts (25%) had severe HCMO, congenital heart disease (CHD) and hydrops fetalis (HF). 2) One fetus with HCMO had glycogen storage disease. 3) One fetus with HCMO had family history of HCMO. 4) Two fetuses had HCMO of unknown etiology. 5) One fetus with supraventricular tachycardia and HF, developed ventricular arrhythmias postnatally; autopsy revealed oncocytic CMO. 6) One fetus had dilated CMO related to maternal hyperthyroidism. 7) One fetus had dilated CMO, and autopsy showed parvovirus myocarditis. Parameters of ventricular function varied, the greater abnormalities were associated with cases with HF (10/31 pts, 32%). Six pts (31%) had severe arrhythmias. Death occurred in 10/31 pts (32%) (1 termination, 2 fetal demise, 7 postnatally).

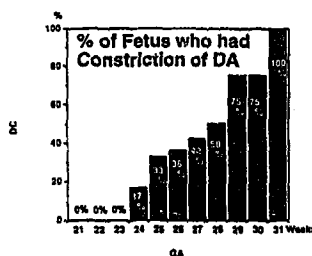
Thus, myocardial diseases are rare in fetal cardiology practice. Its etiology is multiple, most frequently related to maternal diabetes. The association with CHD, arrhythmias or hydrops fetalis has a poor prognosis. Overall survival was 68%.

11:00

746-3 Maternal Administration of Indomethacin and Premature Constriction of the Ductus Arteriosus in the Human Fetus

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Constriction of the fetal ductus arteriosus (DA) may occur during maternal therapy with indomethacin for premature labor. There is, however, little information about the effect of gestational age (GA) or duration of therapy on the constriction of DA. A total 153 Doppler echocardiographic studies were performed on 69 women with 73 fetuses (65 singleton and 4 sets of twins) treated with indomethacin (100–150 mg/day) for premature labor. GA of the fetuses ranged from 21 to 31 weeks. As in previous reports, constriction of DA was defined as Doppler peak systolic velocity > 140 cm/sec and diastolic flow velocity > 40 cm/sec. Constriction of DA occurred in 29 fetuses (39%). No constriction was observed before 23 weeks of GA (Fig). The frequency of constriction of DA increased with gestational age. Constriction of DA occurred within 5 days after starting indomethacin in all. For fetuses ≤ 25 weeks the constriction occurred after a mean of 3.1 ± 1.1 day of indomethacin therapy for those ≥ 26 weeks it occurred earlier, after only 1.6 ± 0.8 days ($p < 0.001$).



Constriction of DA during maternal indomethacin administration for premature labor is observed more commonly after 24 weeks of GA and its frequency increases with advancing age. For fetuses with advanced GA, even a short course of indomethacin therapy may cause DA constriction.

11:15

746-4 Poor Prognosis for Fetuses With Restrictive Foramen Ovale and Left Heart Obstructive Lesions: A Multicenter Study

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It has been reported that restrictive foramen ovale (FO) in infants with hypoplastic left heart syndrome (HLH) decreases the likelihood of success for cardiac transplant or Norwood therapy options. We compared outcomes of 17 fetuses diagnosed between 16–34 weeks of gestation with HLH and 9 with critical aortic stenosis (AS) and dilated left ventricle; in all restriction of the FO was apparent. All fetuses had either a maximal size of the FO ≤ 2.5 mm and/or continuous L → R flow patterns with velocities > 60 cm/sec (up to 2.5 m/sec), or thickening of the atrial septum with no apparent flow. Three fetuses had large, tense L → R bulging atrial septal aneurysms. **Outcome:** Three fetuses died in utero and 4 were electively terminated. Of the 14 with HLH that were born, 2 were not offered surgery and 4 died during attempted stabilization for Norwood surgery or transplant. Four of 6 died after Norwood surgery (preceded in 3 by emergency balloon atrial septostomy), 1 of the 2 transplanted patients survived. Three of the 5 infants born with critical AS died, despite attempts to provide catheter or surgical valvulotomy (4), or transplant (1). The total group yielded only 2 Norwood and one transplant HLH survivor and 2 critical AS survivors (1 balloon, 1 surgical valvuloplasty). The possibility of restricted FO should be specifically investigated on fetal echos with severe left heart lesions and its impact factored into decision-making.

11:30

746-5 Three-Dimensional Reconstruction of Color Doppler Flow Images in Congenital Heart Disease

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We used a 3D reconstruction system designed for viewing structural information for reconstructing in 3D, superimposed color Doppler flow maps of the flows related to regurgitant and/or stenotic lesions or septal defects. Twenty-four patients (1 week–19 yrs) were imaged with an Interspec or VingMed ultrasound system interfaced with a Tomtec computer for 180° rotational transthoracic scan acquisitions. Studies were performed with and without color flow maps superimposed upon the 2D image. Since the 3D system does not currently have separate color processing, color maps were, therefore, selected for discriminating flow, acceleration, aliasing and jets by gray scale. The best 3D images incorporating flow mapping were achieved from discrete, free or wall adherent valvular regurgitant jets using relatively high Nyquist velocities with low color gain. Color Doppler jet geometry, and 3D propagation were well portrayed. Split or dual jets, flow convergences, and jet-wall interactions were also visualized with unique perspectives. 3D flow streams and delineated flow orifices for VSD and ASD shunts provided a unique descriptor of defect size and shunt flow propagation. The addition of color Doppler flow mapping enhanced the 3D echo studies in most patients, and did not significantly increase acquisition or reconstruction time. At times, however, low velocity, broad diastolic inflows obscured tissue information during filling phases. A controller capable of allowing color overlay only during selected gated portions of the cardiac cycle is, therefore, being integrated into the system. Incorporating Doppler flow mapping into 3D imaging can enhance information without compromising structural information and should expand the applicability of 3D methods in pediatric echocardiography.